Clinical Proceedings

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

Contents

Vol. IV **MARCH 1948** **No. 4**

Entered as second class matter November 21, 1946 at the post office at Washington, D. C., under the Act of March 3, 1879. Acceptance for mailing at special rate of postage provided for in Section 538, Act of February 28, 1925, authorized January 17, 1947.

Published monthly by the Children's Hospital, 13th and W Sts., Washington D. C.

Subscription rate is $1.00 per year

DIFFUSE GLOMERULONEPHRITIS

Suggested Classification with Special Reference to So-Called "Lipoid Nephrosis"

Case Report No. 116

Robert O. Warthen, M.D.
John C. Sherburne, M.D.

L. L. 46–1759

L. L., a white male infant was admitted to the Children's Hospital at the age of 20 months because of soft tissue swelling of his eyes, legs, and scrotum of 3 weeks duration. He had apparently been in good health prior to the onset of this illness. A nasal discharge with cough and two days of frequent loose stools accompanied his present illness. He had been fretful and very fussy but his mother believed that due to teething.

The past history revealed that after an uneventful pregnancy the infant was born at term by a normal delivery. He weighed 7 pounds, 8 ounces at birth and was bottle fed without ensuing feeding problems, his diet being adequate in all respects. The patient had experienced exceptionally good health except for an occasional cold and had been previously hospitalized at Children's Hospital at the age of 5 months for a period of 20 days for an upper respiratory infection and an otitis media to which he responded well with penicillin, sulfadiazine and symptomatic treatment. The child had been on a regular diet. He had had no immunizations.

The father, mother and four siblings were all alive and well. The mother had had one miscarriage at six months. A paternal great uncle died of tuberculosis and a paternal uncle had pulmonary tuberculosis. No other familial illnesses or tendencies could be elicited.

The physical examination revealed a 20 month old male who was very fussy and upset on being examined. There was a profuse nasal discharge and the skin was pale, cool and dry. His head was well formed with no abnormalities and the anterior fontanelle was closed. The soft tissues above and below the eyes were so markedly edematous that the eyes were nearly closed. There were some spider-like veins over the anterior chest and the legs, thighs and ankles showed pitting edema. There was a mild injection of the left ear drum with a thickening of the right drum and a purulent exudate draining from the external ear. A profuse serous discharge and severe congestion were noted in both nares. The mouth was clear, the pharynx injected, and the neck supple. The chest was well formed, no abnormalities being noted, and the lungs revealed occasional rhonchi. The heart rate was rapid, the rhythm regular, and the blood pressure 110/70. The abdomen was rounded and palpation revealed the

inferior liver edge to be at the level of the left costal margin. A moderate degree of ascites was present. The reflexes were physiological.

The patient was placed on a high protein, low salt diet and during this admission of 34 days he received 450 cc. of plasma along with 900 cc. of concentrated dried plasma. The diarrhea cleared readily on a protein milk formula. Penicillin and streptomycin were used, but were of doubtful value. The temperature fluctuated during the hospital stay; however, it never exceeded 103° rectally. An electrocardiogram was within normal limits and an x-ray on admission showed an increase in bronchovascular markings suggestive of a bronchitic condition. Kahn and Mazzini tests were negative.

The blood pressure was taken on several occasions, the systolic pressure never exceeding 110 with the exception of an isolated instance of 130, and the diastolic pressure never exceeding 78. The patient's weight was the same on discharge as on admission, i.e. 25 pounds, and during the hospital stay varied between 26 and 22 pounds. The non-protein nitrogen on two samples was 40 and 30 mgm %, and on admission the cholesterol was 335 mgm. %. Serum protein determinations were performed frequently as a guide to the patient's condition. Upon admission the total protein was 2.55% with a serum albumin of 2.21 and a serum globulin of 0.69, or a ratio of 3.2:1; however, this changed to a total protein of 5.21% with a serum albumin of 2.11, a serum globulin of 3.10 and a ratio of 0.68:1. Blood studies revealed a hemoglobin ranging from 10 to 13.5 gms. The red blood count improved from 3.5 to 4.9 million before his discharge and the white blood cell counts were all within normal limits as were the differentials, with the exception of an eosinophilia of 8% on admission. Blood smears showed very few abnormal red blood cells. Urinalyses always revealed large amounts of albumin. On admission 2,800 mgm. were noted; however, during the hospital stay the readings usually varied from 2000 to 5000 mgm., although an occasional finding as low as 60 mgm. was recorded. Most of the microscopic urine examinations revealed many variations of hyaline and finely granular casts. Several samples showed white blood cells and one examination out of thirty showed "a rare red blood cell."

After a somewhat stormy hospital course he clinically improved and it was deemed advisable to transfer him to a convalescent home for special diet and care.

During the brief stay at the Children's Country Home his fluctuating edema and ascites persisted and after 14 days in the home he was readmitted to this hospital because of a temperature of 102° and lethargy of one day duration.

At the time of this readmission, physical examination revealed a 21 month old, fairly well developed, fairly well nourished, moderately edematous

white male infant appearing chronically ill. His temperature was 103° rectally. The skin was pale and hot, the upper eyelids were moderately puffy and there was an acute bilateral catarrhal otitis media with an old greyish white scar in the posterior inferior quadrant of the left drum. The mucous membranes were pale and the nasopharynx was moderately injected as were the hypertrophied tonsils. Bilateral cervical lymphadenopathy was noted and the neck was freely movable. The heart rate was 190 per minute, P2 was greater than A2 and a systolic murmur was heard with greater intensity over the pulmonic than the mitral area. The heart was not enlarged and the blood pressure was 128 over 84. The lungs were clear to percussion and auscultation, respirations being 20 per minute. The abdomen was distended and rotund, and the umbilicus was moderately herniated and edematous. A fluid wave could be elicited though the liver and spleen could not be palpated due to severe abdominal tenderness and distention, the liver was percussed to 2 cm. below the right costal margin. No abdominal tumors or masses were observed. There was one plus pitting edema of the scrotum, while the penis was only mildly edematous.

During this admission he developed a slight case of diarrhea which lasted several days. Penicillin therapy and supportive measures were employed for 5 days with a resultant clearance of the otitis media and diarrhea. The edema and the ascites were treated with dried plasma on several occasions with very little persistent response. The albumin in the urine varied from 2000 to 800 mgm. on numerous occasions. Many white blood cells, a moderate number of fine and course granular casts and occasional red blood cells per hpf were reported during this stay. The non-protein nitrogen was 28 and 34 mgm % and the total proteins were 2 83% with a ratio of 4.5:1. On admission a leucocytosis of 16,500 was present with a slight shift to the left; however, the leucocytes returned to normal within a few days. There was a mild anemia of 3.1 to 3.8 million red blood cells and a hemoglobin of 10 grams. The patient appeared somewhat improved after about two weeks of hospitalization and was again discharged to the convalescent home.

He was again admitted here for another period of hospitalization at the age of 24 months with the following history. On the day before this admission the patient played and chattered as in the past and had eaten his meals well. About midnight he awoke and appeared very fretful and restless, this episode persisting for over two hours. At that time his temperature was 102° and five hours later rose to 104.8°. Aspirin and symptomatic measures were employed and because of a poor response he was again returned to this hospital.

At this time as before, he appeared chronically ill with generalized edema

and a tense protuberant abdomen. His edema had improved somewhat since his discharge; however, his mucous membranes remained pale and his right ear was slightly injected as was his pharynx. The neck was supple. Throughout the chest were scattered fine moist rales. The heart was essentially normal. A left inguinal hernia was readily reduced leaving a cystic mass in the region of the testicle. The extremities and the neurological were negative as was an admission chest x-ray.

Concentrated dried plasma was given following which more edema appeared. This was followed by albumin after which the edema promptly receded only to return within a few days. Paracentesis was performed at that time and was repeated several days later.

The total protein was 2.41% with an A/G ratio of 0.5:1 on admission and increased to 4.55% with an A/G ratio of 0.9:1, only to drop to 3.73% with an A/G ratio of 0.6:1 later. Urinalyses always showed large amounts of albumin, never exceeding 5000 mgm. or dropping below 1000 mgm. As before the microscopic examination on 22 specimens showed, as a rule, moderate numbers of hyaline casts, some coarse and fine granular casts and few to moderate numbers of white blood cells. Three samples had "rare red blood cells," while two had a "few red blood cells." On admission a mild leucocytosis was found but this was only temporary. An anemia present on admission improved to four million red blood cells and 12.5 grams of hemoglobin upon discharge.

After 44 days of hospitalization the patient had improved sufficiently, with the aid of penicillin and supportive measures, to convalesce at home.

Though he was apparently improved upon discharge he soon developed "diarrhea" and a "swollen abdomen." His stools became yellowish and moderately soft numbering two to three per day; however, he received no medication and was placed on a salt free diet. He progressed fairly well until the day before his final admission, nine days after his previous discharge, when his mother noted his stools becoming looser and more frequent. Shortly thereafter he developed abdominal pain accompanied by a "grasping" of the umbilical region and "crying out." Marked restlessness followed and the severe pain in the abdomen continued causing the child to "grunt" frequently. The stools became more frequent and watery and the child vomited several times. Again he was hospitalized.

The final admission physical examination revealed an acutely ill 25 month old white male child with grunting respirations, extreme pallor and a markedly distended abdomen. There was no nuchal rigidity and the fontanelles were closed. The mucous membranes were extremely pale and the lips were dry. The lungs were clear to percussion and auscultation and the heart rate was rapid, no murmurs being heard. The abdomen was distended with a large umbilical hernia, the spleen was not palpable

and the liver was enlarged to two finger breadths below the right costal margin. There was pronounced dulness in the flanks. The abdomen was tender and doughy and over its upper portion were prominent dilated veins. The genitalia were normal for a male of two years with the exception of a large left inguinal hernia. There was no edema of the extremities at this time.

The diarrhea and apparent acidosis on admission were treated with intravenous lactate and plasma as well as penicillin; however, the temperature remained elevated between 103° and 104° rectally and very little improvement was noted. Two carbon dioxide combinings power examinations were reported as 35 volumes per cent and 17 volumes per cent, in that order, and the total protein was 3.60%. The leucocyte count was 9,400 with a normal neutrophile count. Labored respirations continued throughout the hospital stay and his general condition suddenly became worse with circulatory collapse and death supervening on the second day of hospitalization.

NECROSPY REPORT

The body was that of a poorly nourished, pale white male infant appearing the stated age of two years. There was a thick yellow fluid exuding from the nostrils, distended abdomen and what appeared to be a large umbilical hernia. The scrotum was discolored and markedly swollen, particularly on the left side.

On section the skin was pale and thin and there was very little subcutaneous fat. The pleural cavities contained approximately 10 cc. of clear yellow fluid (culture revealed non-hemolytic E. coli). The peritoneal cavity was distended with 150–200 cc. of clear to cloudy yellow fluid (culture revealed pneumococci and non-hemolytic E. coli). The umbilical sac contained a thick yellowish-green purulent exudate which was easily expressed through the ring into the peritoneal cavity. The superior surface of the liver and the inferior surface of the diaphragm were finely adherent and were coated with a thin, green, filmy exudate. The stomach, colon and spleen were densely adherent and the spleen was covered by a similar greenish-yellow exudate. The remainder of the intestinal loops were not significantly affected by the inflammatory exudate.

The left internal inguinal ring was open widely, measuring 4–5 mm. in diameter, and admitted a large probe easily to the inferior wall of the scrotum. No abdominal contents were herniated through the ring; however, the scrotal sac contained a thick greenish-yellow exudate similar to that in the peritoneal cavity.

The pericardium contained a small amount of clear yellow fluid and there were no adhesions. The heart was not enlarged and the valvular cir-

cumferences were within normal limits. The right ventricular wall was firm and measured 3 mm. in thickness and the left was flabby and measured 10 mm., the right chambers being somewhat dilated.

There was an aberrant renal vein arising in the upper pole of the right kidney and draining into the inferior vena cava above the entrance of the renal vein.

The trachea revealed no abnormalities and the lumina of the main bronchi were filled with a thick tenacious yellow fluid. The lungs disclosed gross congestion and edema, the congestion being more marked at the bases.

The liver weighed 498 gm. (normal 394 gm.) and extended 4 cm. below the right costal margin in the mid-clavicular line. The external surface has been previously described and the section surfaces were somewhat congested with an occasional pale yellow-white patch.

The spleen weighed 66 gm. (normal 33 gm.), measured 9 x 5 cm. and was enlarged and coated by a filmy greenish exudate. On the section surface the follicles were obscured by a soft, dark red pulp which scraped with ease.

The small intestine was slightly distended and the walls of the small and large intestine were thickened with hypertrophied lymphatic tissue. The mucosa was pale and in some parts swollen. There were no visible rupture points.

The right kidney weighed 88.5 gm. (normal 47 gm.) and the left 82.7 gm. (normal 46 gm.). The perirenal fascia were markedly thickened and adherent to the colon. The renal capsules stripped easily revealing smooth, pale glistening cortical surfaces. The kidneys were enlarged, pale and soft and on section the parenchyma was pale, slightly swollen and "greasy" to the touch. The cortex was pale, contained a few whitish specks bordering the pyramids and was well differentiated from the medulla. The pyramids were dark and red.

Coronal sections of the brain showed marked congestion and there was suggestion of atrophy of the anterio-frontal gyri. The pons and cerebellum also were congested.

The remainder of the gross examination was essentially negative.

Clinical Diagnosis:

1. Lipoid Nephrosis
2. Peritonitis

Pathologic Diagnosis:

1. Peritonitis due to pneumococcus, with
   Suppurative hydrocoele, right
   Suppurative oomphalitis

2. Subacute and chronic glomerular nephritis, proliferative (nephrotic type)
3. Pulmonary edema and congestion.
4. Umbilical hernia.

Microscopic examination of the tissues confirmed the pathological diagnosis; however, for the sake of discussion the microscopic kidney findings will be presented in detail.

Many of the glomeruli disclosed marked proliferation of the epithelial and endothelial cells with hyaline degenerative changes of the latter and of the capillary loops. Slightly thickened basement membranes were prominent. A moderate number of glomeruli completely filled Bowman's capsules and adhesions between the capsules and capillary loops were evident. A rare capsule contained a thin layer of proliferated epithelial cells suggestive of a crescent formation and a moderate number of the glomeruli were relatively bloodless.

Many of the tubules contained albuminous casts and the lining cells revealed varying stages of albuminous degeneration. Lipoid degenerative changes were evident in the collecting tubules.

DISCUSSION

Many pathologists have ceased to consider lipoid nephrosis as a separate clinical entity, believing that it is a form of glomerulonephritis. With that viewpoint in mind this paper was written, for the case here presented was a clear cut one of lipoid nephrosis prior to death but the postmortem examination revealed a picture of chronic (proliferative) glomerulonephritis.

In order to develop our explanation of the pathogenesis of so-called "lipoid nephrosis," which we choose to term chronic proliferative glomerulonephritis, it seems advisable to review first the microscopic pathological findings in glomerulonephritis and relate them to the case here presented.

As is generally known, there are two forms of diffuse glomerulonephritis, proliferative and exudative. The acute proliferative type is pathologically characterized by a proliferation and swelling of the endothelial cells lining the glomerular capillary tufts and a thickening of the basement membrane producing relatively bloodless tufts. There is also a proliferation of the epithelial cells lining the tufts and capsule, with a desquamation of the tuft cells into the capsular space. In the subacute or chronic proliferative type, fibrosis and minimal to moderate hyalinization of the tuft endothelial cells predominates with an occasional case of fibrous adhesions between the tuft and capsule producing a subdivision of the glomerular space into crypts, thus giving the appearance of a lobulated glomerulus. Because of these changes the tufts are relatively bloodless. Also an occasional thin

epithelial crescent-like formation may be present due to hyalinization of the proliferated capsular cells and the desquamated tuft cells. Due to the minimal hyalinization these damaged glomeruli do not contract as do those of the exudative type of glomerulonephritis to be described below and for that reason the tuft capillaries are only partially occluded. This microscopic picture of the glomeruli of subacute or chronic proliferative nephritis is similar to that of the case presented and is also similar to that described in the literature as "lipoid nephrosis."

Clinically the proliferative form is often overlooked in the acute stage due to the mildness of the pathological condition as well as the lack of an exudate such as is seen in the acute exudative form. The subacute or chronic proliferative stage, which we believe to be so-called "lipoid nephrosis," shows minimal to moderate hyalinization of the glomerular tufts and is clinically evidenced by albuminuria, marked edema, hypercholesterolemia, low plasma proteins and a reversal of the A/G ratio. This is explained on the basis of a partial obstruction of the glomerular tufts, this partial vascular bed occlusion allowing the continued urinary escape of albumin in large quantities.

Acute exudative (hemorrhagic) glomerulonephritis is pathologically characterized by a picture similar to that of the proliferative form with an additional more marked proliferation of the epithelial cells lining the glomerular tufts and Bowman's capsule plus a purulent capsular exudate composed of leukocytes, red blood cells, epithelial cells and fibrin. Due to the more severe damage of the tuft the leukocytes and red blood cells invade the tuft and are cast off into the intracapsular space. In the subacute exudative form one finds epithelial crescents which consist of capular exudates that have been organized and in the chronic phase the picture of varying stages of hyalinization and contraction of the crescents and glomerular tufts predominates, finally resulting in bloodless glomeruli.

Clinically the acute exudative stage produces more marked findings than the proliferative stage due to the greater degree of glomerular damage, hence the frequent occurrence of red blood cells and white blood cells in the urine. This clinical entity of acute exudative or hemorrhagic glomerulonephritis is well known and will not be discussed here. In the subacute or nephrotic stage of exudative glomerulonephritis the clinical picture may be identical with that of subacute or chronic proliferative nephritis or so-called "lipoid nephrosis"; however, due to the more severe damage and the varying stages of contraction of the tufts there may be an associated hypertension, elevated non-protein nitrogen and urinary red blood cells. When this stage progresses to the chronic phase these findings become more marked because of the complete hyalinization and occlusion of the tuft vascular bed and terminably only scant amounts of urine pass through the tuft.

In both conditions previously discussed there are secondary vascular changes producing degenerative kidney tubular damage; however, we believe these tubular changes to be of secondary importance in arriving at a microscopic diagnosis.

The pathological findings discussed in this paper are nothing new in the literature, having been recognized for many years. This case, as may have been gathered, is presented to offer an explanation of the pathogenesis of so-called "lipoid nephrosis." As we have mentioned and attempted to explain in an orderly pathogenic fashion, lipoid nephrosis is merely a subacute or chronic stage of proliferative glomerulonephritis and is probably etiologically similar to exudative nephritis, the chronicity and severity in either instance depending on the number of glomeruli damaged and the extent to which they are damaged. We might also postulate a possible explanation for complete cures of "lipoid nephrosis." These may be due to a complete obliteration of the damaged leaking glomeruli of which there may be few in number and the intact remaining glomeruli may be sufficient for normal function.

In summary, it becomes evident that pathologically the *amount* of glomerular damage of subacute or chronic proliferative nephritis (lipoid nephrosis) is comparable to that of subacute exudative glomerulonephritis, both conditions being marked by partially occluded tufts, hence the clinical resemblance of the two conditions; however, subacute exudative glomerulonephritis may be involved with a more severe form of damage with resultant coexisting contracture of many glomeruli producing completely occluded tufts and associated clinical findings of chronic exudative glomerulonephritis.

A suggested future classification of diffuse nephritis follows, with the corresponding present day terminology in parentheses:

1. Proliferative glomerulonephritis
   a. Acute (none)
   b. Subacute—chronic (lipoid nephrosis; nephrosis)
2. Exudative glomerulonephritis
   a. Acute (acute hemorrhagic nephritis; acute glomerular nephritis; acute exudative nephritis)
   b. Subacute—chronic (glomerular nephritis with nephrotic component; subacute glomerular nephritis; chronic glomerular nephritis)

MESENTERIC VASCULAR OCCLUSION IN CHILDREN

With Report of a Case

Case Report No. 117

Howard S. Madigan, M.D. B. L. 47–13429

Occlusion of the major mesenteric vessels in children is rare. Recognition of the condition clinically is almost never achieved. The paucity of literature and reported cases prompts this writing, to review this entity and to report a case which occurred following intussusception.

REPORT OF CASE

B. L., a 7-month-old white female was admitted in the early afternoon, on December 26, 1947. The infant had been fretful for one week, and intermittently seemed to have abdominal discomfort. Nothing definitely abnormal about the stools had been noted. On the day prior to admission, a blood-streaked mucous stool was passed. She vomited several times during the night. An enema was given on the morning of the day of admission, the returns consisting of clotted bloody mucous.

The birth and previous history were essentially unrevealing. The infant had been in good health prior to the onset of the present illness.

Examination on admission revealed an acutely ill infant with a temperature of 99.2°. The abdomen was distended and there was marked splinting on the left side. Rectal examination was done and an indefinite mass, high on the left side was palpated. There was bloody material on the examining finger.

A diagnosis of intussusception was made and operation performed about three hours after admission. The intussusception was reduced without too much difficulty. The involved bowel was markedly edematous, but of good color. The appendix was also removed (it had been included in the intussuception).

Immediately post-operatively the infant seemed in fairly good condition. About four hours after operation, however, the temperature rose to 106° (per rectum), she became pale, but not cyanotic. Attempted administration of intravenous fluids was unsuccessful. Within 3 to 4 hours, the respirations became rapid and shallow and her condition became extremely poor. Oxygen therapy was of no avail, and the infant expired approximately eight hours post-operatively.

Significant findings at necropsy were confined to the abdomen. The gastrointestinal tract was patent throughout. From a point 7.0 cm. proximal to the ileocecal valve to the splenic flexure, the bowel was hemorrhagic

and purple in color. The superior mesenteric artery and vein were thrombosed. There were a number of enlarged mesenteric lymph nodes in the region of the ileocecal valve. The appendix, when examined grossly, was firm, and the external surface appeared hemorrhagic. The distal part was friable.

DISCUSSION

The first case of mesenteric vascular occlusion was reported by Tiedeman in 1843.[1] Accumulation of series of cases was slow. Trotter, in 1904, reported on 308 cases, only 13 of which were diagnosed pre-operatively or before autopsy. Four of these cases occurred in children, the youngest of whom was five years old. The next sizeable series, 214 cases, was reported by Jackson, Porter and Quinby[2] in 1914. No specific mention is made of the age incidence. Sporadic reports of cases occurring in children are found in the literature.[3, 4, 5, 6, 7]

Johnson[1], in 1940, reviewed the English literature, and could find only 11 cases of mesenteric vascular occlusion in children, to which he added his own case report. In 1942, Laufman and Scheinberg[8] reported forty-four cases, three of which were in children. Two were infants: an 11-day old infant developed an occlusion following the accidental sectioning of the superior mesenteric artery during an operation for the release of a congenital duodenal band; a 10-day old infant had an acute omphalitis, with thrombosis of the umbilical, splenic and portal veins, and died of peritonitis and sepsis. The mother had puerperal sepsis.

Incidence and Etiology

As mentioned, the condition is extremely rare in children. In reported cases, males are affected more often, the ratio being about 2:1. The age range is from the first few days of life throughout childhood, but most cases have been in those under 10 years of age.

The etiological factors are similar in the young and the aged. However, infection and heart disease play a greater role as inciting factors in the young. Warren and Eberhard[9] have placed the etiological factors into four categories, as follows:

(1) Known infection, e.g., appendicitis, thrombophlebitis, pelvic abscesses, peritonitis and general sepsis.

(2) Hematogenous causation: blood dyscrasias or changes known to predispose to thrombosis.

(3) Traumatic: trauma of any sort to the mesenteric vessels; tearing of the mesentery and trauma from abdominal operations.

(4) Mechanical (the largest group): portal stasis, pressure from tumors, pressure from adhesions or congenital bands.

A possible etiological factor has been advanced in patients undergoing ablation of the lumbar sympathetic chain as a therapeutic measure. Two cases (in adults) have been reported[8] in which mesenteric thrombosis occurred following sympathectomy. Sufficient evidence has not been accumulated to recognize this as an established causative factor.

The traumatic and mechanical disturbances incited by intussusception are of etiologic significance in occlusion of mesenteric vessels in children. However, this complication is extremely uncommon. In a study of ninety-two cases of intussusception, Gibbs and Sutton[10] did not report mesenteric vascular occlusion among the complications.

Physiological and Pathological Considerations

The occlusion may be arterial or venous, as the result of embolism or thrombosis. Embolic arterial occlusion is most common, and the superior mesenteric artery is most often involved.[11] This latter fact is based on the early exit of this artery from the aorta, and the fact that it is a more direct continuation of the aorta. Venous occlusion is almost always due to thrombosis. It is usually associated with infection in the abdominal organs tributary to the portal vein. Appendicitis, pelvic infection and strangulated bowel are frequent precursors.

The results of occlusion of mesenteric vessels depend on several factors:[8] (1) whether the occlusion is arterial or venous; (2) the caliber of the vessel involved; (3) the suddenness of the occlusion; (4) the length of time the occlusion persists before operation or death; (5) the length of intestine affected by the occlusion.

The superior mesenteric artery, though not an end-artery, behaves like one. Sudden occlusion results first in a violent contraction of the intestine from the ligament of Treitz to the middle of the transverse colon. This is interpreted as an anoxic response to the sudden cessation of arterial blood flow. The bowel is firm, white, and rippled as a result of the contraction of both muscular coats. The intramural vessels are compressed and the capillary blood is drained off by the veins. As the intestinal musculature becomes fatigued (or toxic), it loses its contractability and relaxes. Some parts relax more quickly than others, so that after a few hours the bowel has a mottled appearance, bluish areas appearing between blanched areas. After about eight hours, the entire involved bowel becomes relaxed and is blood-filled.

Results of experimental investigation show that the mechanism of hemorrhagic infarction following arterial occlusion is due partly to regurgitant flow from the veins and partly to reflux of arterial blood through collateral channels. As the bowel musculature relaxes, a negative pressure is created in both arteries and veins sufficient to draw blood back into the intestinal wall. This will occur even if the artery and vein are ligated.

Sudden occlusion of the superior mesenteric vein (experimental) shows that there is an initial bluish discoloration of the bowel because of relaxation rather than contraction of the intestinal musculature. Blood volume is decreased, blood pressure falls, and death ensues rather rapidly.

Such sudden venous occlusion is uncommon clinically, the process being more gradual. When a thrombus forms slowly, there is time for the development of collateral circulation. With such a gradual occlusion, there may be intestinal infarction or there may be minimal or no evidences of such a disturbance. Experimentally, gradual occlusion results in death after a period of time from chronic anoxia. Occlusion of the secondary mesenteric vessels causes no circulatory embarrassment of the intestine.

Clinically, where a combination of arterial and venous occlusion occurs, the end results are variable, and the severity of the manifestations as well as the suddenness of death are governed by the predominance of one or another of the factors discussed.

When a shorter segment of bowel is involved, blood loss plays a less important role and the deprived bowel wall becomes ready ground for bacterial invasion. Thus gangrene is more apt to be found if a short segment is involved, and death occurs from perforation and peritonitis. If depletion of the blood volume takes place rapidly, death supervenes before gangrene can manifest itself. The duration of the occlusion is not the sole factor in the production of gangrene.

Experimentally, it has been concluded that if more than 5.0 cm. of intestine be deprived of arterial supply, the segment will undergo infarction and gangrene. In shorter segments, the circulation is preserved at each end through the intramural channels. A somewhat longer segment can survive after venous occlusion alone than after arterial occlusion alone.

Pathologically,[12] the affected portion is thickened and dark red in color. The limits of gangrene are usually sharply defined, but the demarcation may be more gradual. The serosa is covered with an inflammatory exudate. The bowel wall is filled with blood, and the mucosa is necrotic and may be ulcerated. The lumen usually contains thick tarry blood. The mesentery is thickened and contains large hemorrhagic patches. The mesenteric veins are engorged and the mesenteric glands are swollen and hemorrhagic. There is a variable amount of blood fluid in the peritoneal cavity or there may be a general peritonitis. There will be thrombosis of the major mesenteric vessels involved.

Clinical Features

Because mesenteric vascular occlusion as a primary condition occurs so infrequently in children, a description of the clinical aspects is difficult. Failure to recognize the condition cannot be construed as faulty diagnostic acumen. However, a knowledge of the clinical manifestations is of value

in the differential diagnosis of acute abdominal disorders in young individuals. It must be realized that in children, as well as in adults, the manifestations of vascular occlusion are most often superimposed on some reasonably well-established clinical entity.

Insofar as symptomatology is concerned, intestinal obstruction, strangulation or rupture of a viscus may be simulated. Pain is usually quite sudden in onset, persistent and agonizing. Vomiting is a constant feature and may even overshadow the pain. Not infrequently the vomitus may be bloody. Melena or occult blood in the stool often occurs when the occlusion is a venous one. Varying degrees of shock are present, being particularly evident when occlusion is followed by infarction. The temperature is usually normal or subnormal at the onset, but becomes septic in a matter of hours. Abdominal tenderness is usually moderate as compared with the severity of the pain. This tenderness is generalized; at times shifting tenderness can be elicited. Distention and rigidity of marked degree are not common. A leucocytosis of from 15,000 to 30,000 may be encountered, unless the process is fulminating, in which event no rise in the white-cell count will be evident.

In children, particularly, it is difficult to determine the time of onset, primarily because of the usual presence of some predisposing or exciting cause. There is no apparent relationship between the duration of symptoms and the severity of the lesion. As a rule, symptoms are of longer duration in cases of venous occlusion.

Treatment

The general condition of the patient must be accorded prime consideration. Shock should be treated before any surgical measures are considered. Transfusions of whole blood in substantial amounts is needed to restore blood volume. Successful operative intervention in children has not been reported. This is not surprising if consideration is given to the poor condition which such patients present.

If the diagnosis be made and surgery done, the type and extent of such procedures will be governed by the amount and degree of bowel involvement. Ideally, resection of the involved segment is the procedure of choice. Anti-coagulant therapy post-operatively is advised to reduce the possibility of further thrombotic accidents. If no operative procedures are carried out, it is dangerous to administer anti-coagulants, for added blood and fluid will be lost into the lumen of the bowel and into the peritoneal cavity.

Prognosis and Mortality

No cases of recovery in children have been reported. The establishment of an adequate collateral circulation is a remote possibility. Death in

cases with venous occlusion without infarction is due to sepsis, toxemia or liver insufficiency.

If the diagnosis is made, and operation not performed, the mortality is virtually 100%. Considering all cases of mesenteric vascular occlusion, Whittaker and Pemberton[13] report a mortality of 74% in their series. In statistically significant series, a mortality rate of 60% is the lowest reported.

SUMMARY

A case of mesenteric vascular occlusion following intussusception is reported.

Mesenteric vascular occlusion in children is a rare condition, and is almost never diagnosed clinically.

A review of the literature and a discussion of the clinical entity is presented.

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STREPTOCOCCAL MENINGITIS

Report of a Case Complicating Pertussis

Case Report No. 118

Allan B. Coleman, M.D. B. S. 47–8810

Meningitis is a relatively infrequent complication of pertussis according to Sauer.[1] Although previously a common disease in children, meningitis caused by the hemolytic Streptococcus has become uncommon in the ten years which have elapsed since the introduction of the sulfonamide drugs into clinical usage. In this hospital, Streptococcal meningitis has comprised less than 1% of all cases of purulent meningitis seen in the past five years.[2] The following case, therefore, is presented as a clinical rarity.

A two year old white girl was admitted to the Children's Hospital because of cough, fever and vomiting. A persistent cough began six weeks prior to admission; two weeks later the cough became paroxysmal and was often followed by "whooping" and vomiting. Three older siblings developed similar symptoms concurrently. Fever was first noted two days before admission, but the cough seemed to have decreased in severity. Shortly before admission a red area appeared on one buttock.

The patient was born at home in a nearby city. The birth, feeding, and early development were normal. There were no previous serious illnesses. No immunization had been administered. The family history was non-contributory.

Physical examination revealed a well developed and nourished two year old, appearing semi-comatose, moderately cyanotic and obviously seriously ill. There was a small vesicle on the right buttock; surrounding this lesion was a palm-size, hot, red, indurated area. The mucous membranes appeared dehydrated and the pharynx was diffusely injected. The lungs showed generally diminished resonance, harsh breath sounds and scattered moist rales bilaterally. The neck was rigid and Kernig and Brudzinski signs were present. The plantar reflexes were extensor bilaterally. The deep reflexes were sluggish.

Lumbar puncture revealed thick, milky spinal fluid under increased pressure containing 300 mgm. % of protein, no sugar, and 32,000 leukocytes per cubic millimeter, with 76% polymorphonuclear cells, 18% lymphocytes and 6% endothelial cells. Direct smear of this fluid contained many cocci staining "gram-variable."

Other initial laboratory studies revealed a hemoglobin of 12 grams, 4.5 million erythrocytes and 26,000 leukocytes with 56% segmented neutro-

philes and 8% stab forms. The urine was concentrated and contained 100 mg. of albumin per 100 cc. and a few hyaline casts and leukocytes. The carbon dioxide combining power was 54 volumes per cent. A blood culture was sterile. A roentgenogram of the chest showed a localized area of bronchopneumonia on the right.

At the time of the initial lumbar puncture, 10,000 units of penicillin was administered intrathecally. Sodium sulfadiazine in dosage of two grains (0.13 gm.) per pound of body weight was given subcutaneously in divided doses at 8 hour intervals. The blood sulfadiazine level was 18 mgm. per

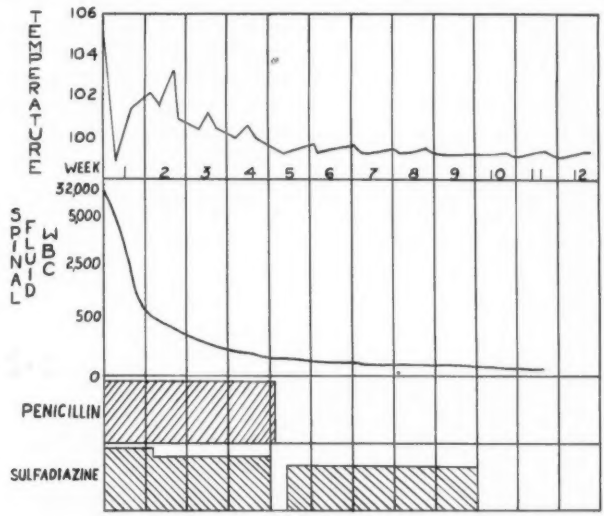

FIG. 1

100 cc. the morning of the first hospital day. At that time cultures of the spinal fluid showed a hemolytic streptococcus, with a penicillin sensitivity of 0.0156 unit of penicillin per cubic centimeter.

Penicillin was begun intramuscularly in doses of 50,000 units every 3 hours and continued until the 29th hospital day. Supportive measures included oxygen, parenteral fluids, blood transfusions and 40 cc. of human hyperimmune pertussis antiserum in divided doses intramuscularly.

The patient received a total of 7 intrathecal installations of penicillin consisting of 10,000 units in 10 cc. of physiological saline during the first nine hospital days.

The temperature reached normal during the second hospital day (see figure) but rose again to 101°F. It remained elevated between 100° and

103° during the next 19 days, falling by slow lysis to normal on the 21st days. The temperature remained normal during the remainder of the hospital course.

The spinal fluid showed a rapid reduction in leukocyte count (see figure), reaching 2,900 on the third day. Sugar appeared in the spinal fluid on the 5th day. No organisms could be recovered from the spinal fluid after the third day at which time the patient was conscious and began to take fluids and medication by mouth. The inflammatory lesion of the buttock healed rapidly. The pneumonic process resolved in a few days.

Sulfadiazine was discontinued on the 30th day and begun again on the 33rd day because of persistence of 260 leukocytes and 200 mgm. % of protein in the spinal fluid. Sulfadiazine in dosage of one grain (0.06 gm.) per pound of body weight daily was continued until the 63rd hospital day. During this time no evidence of localized brain disease could be elicited and roentgenographic studies of the mastoids and paranasal sinuses were negative on two occasions. The spinal fluid leukocyte count fell slowly reaching 30 per cubic millimeter on the 75th hospital day.

The patient was discharged on the 83rd hospital day in good condition. She was last seen approximately 4½ months after her hospital admission at which time the spinal fluid was normal and there were no apparent residduals of her illness.

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SOME NOTES ON ERYTHROBLASTOSIS FETALIS

Harry C. Robelen, Jr., M.D.

From January 1944 through September 1947 there were 66 patients diagnosed as having Erythroblastosis Fetalis in this hospital. After a preliminary study of the case histories, it was deemed advisable to establish a criterion for diagnosis in order to put the review on as accurate a basis as possible. Therefore only those cases in which an antibody titer on the mother or baby was recorded or unless the postmortem findings were compatible with the diagnosis were accepted. This reduced the total number by 39 cases, leaving 28 cases for consideration.

According to Potter, available data indicates that not more than 3 to 4% of Rh negative women produce antibodies which injure their offspring. If antibodies have not been produced by the fifth pregnancy, the likelihood that they will subsequently form is lessened somewhat. When the husband is heterozygous Rh positive, at least 50% of the infants will be Rh negative and free from trouble.

Of the 66 total cases, 15 died. Eight of these were excluded from this review because postmortem examination failed to produce any concrete evidence of erythroblastosis.

Of the 28 cases studied there were:

	Recovered	*Died*
White:		
Male	12	5
Female	7	3
Colored:		
Male	1	0
Female	0	0
	20	8

Twenty-five cases had received one or more Rh negative transfusion. Three had received mixed Rh positive and negative blood; the remainder had received none. The use of Rh negative blood is generally accepted on the basis that the Rh negative cells of the infant are being destroyed by the maternal antibodies that pass into the fetal circulation and that any additional Rh positive cells introduced by transfusions will be affected also. There are some writers who advocate giving Rh positive cells in order to remove more quickly the circulating antibodies from the fetal circulation but this opinion is in the minority.

Breast feeding is contra-indicated since the maternal antibodies are known to be transmitted via the breast milk and to be absorbed intact by

the infant. Two of our cases received replacement transfusions and their cases are as follows:

Case # 1: Labor was induced at 33 weeks because of a rise in anti-Rh titer to 1:64 with incomplete or blocking antibodies also present. The delivery of a white male infant was uneventful. Two days later the infant became slightly jaundiced and was admitted to Children's Hospital. The red blood cell count on admission was 5 million with 2 erythroblasts per 100 white blood cells.

An exsanguination-replacement transfusion was done the same day with 400 cc. of fresh citrated Rh negative blood given into an ankle vein while 350 cc. of blood was removed simultaneously from the radial artery. The procedure required 50 minutes during which time 0.4 cc. of 1–1000 heparin was given. His immediate condition was described as good but he died 5 hours after the transfusion was completed.

Blood removed from the infant showed the following:

	1st 50 cc. removed	*last 50 cc. removed*
Icterus index	200 u	150 u
Total protein	5.86%	4.48%
Rh factor	Positive	Negative

Case # 2: A white male was transferred from Georgetown Hospital at 4 days of age. The labor was induced, and an exsanguination-replacement transfusion performed 15 minutes after delivery. Five hundred cubic centimeters of Rh negative blood was given via the umbilical vein, while 350 cc. of blood was withdrawn from a radial artery of the infant. He was given 100 cc. of Rh negative blood in this hospital 4 weeks later and recovery was uneventful.

In a personal communication, Davidsohn states, "The treatment of choice at the present time is no doubt the so-called replacement or exchange transfusion. There is of course the necessity of determining the proper indication for transfusion. One must be sure that the child actually had fetal erythroblastosis and that is not always apparent at first. On the other hand, waiting until the clinical manifestations become apparent may be too long. We are guided by the obstetrical history and by the studies of Rh antibodies during pregnancy. We determine the Rh factor of the baby immediately after birth, also the blood count and the so-called Coomb's test. We are guided mainly by the Coomb's test and the presence of Rh incompatibility. The introduction of the blood into the baby is a relatively simple matter. It is the removal of blood from the baby that presents some difficulty. My personal experience has been limited to the umbilical vein for which a special plastic catheter can be used. We have treated a small number of babies by this procedure and found it rather

satisfactory. In giving replacement transfusions, one may give it rather rapidly because of the withdrawal of the blood which removes the burden on the child's circulation. It takes us approximately one to two hours to do the whole procedure. The results in some instances have been nothing short of miraculous." He also states that mothers are not permitted to nurse the babies.

Wiener in another communication refers to the spectacular effect of exchange transfusions. His technique entails the use of heparin, injection of the donors' blood into a vein at the ankle and the withdrawal of the patient's blood through the radial artery at the wrist. Of a total of seventeen infants treated by this technique, many of whom were critically ill, all but one made a prompt and complete recovery. Wiener considers the exsanguination replacement method an effective and safe procedure.

An interesting historical sidelight is the report of the first exchange transfusion on an erythroblastotic infant in 1925 by Hart "to remove enough toxins from the blood to prevent progress of the disease." The patient recovered but the technique was apparently dropped until recent years.

Levine states "It is my policy to recommend replacement transfusion only where the mother has been intensively immunized and gives a history of a previous transfusion of Rh positive blood or of having had an erythroblastotic infant. In the absence of these two conditions, replacement transfusion may be carried out in those cases where the mother had potent antibodies which appear early in the last trimester. In those cases in which the antibodies appear only in the last weeks of the pregnancy, the infant may be either entirely normal or have a very mild form of the disease. Even under these conditions I have had occasional disappointing results due to the lack of correlation of severity of symptoms and serologic findings. On the whole, however, the rules given above will apply. I had forgotten to mention additional tests which may also serve as a guide and they are the serologic tests on the cord blood at delivery. If the infant's cells are completely coated with blocking antibodies so that the blood behaves as though it were Rh negative, such cells will be found to give strong reactions with the anti-human globulin serums. In my opinion this is always an indication for a replacement transfusion although I have seen such infants recuperate with numerous single transfusions. If however, the washed cord cells give a comparatively weak reaction, such infants may be treated more conservatively."

An attempt was made to obtain a follow-up on as many of the total number of patients as possible. Of the 51 living, 15 were contacted and 9 were brought in for additional blood examinations with as many of the immediate family as possible. The result is shown in the following charts.

1. J. H. August 1944. No previous titer

Rh	Mother	Father	10 yr.	7 yr.	5½ yr.	3 yr. (Pt.)
−	+	+	+	+	+	+ living & well

Screen test Agglutination slight with 2 Rh positive bloods.
In albumin 1:1
In saline 0
Incomplete or blocking 1:1

2. M. S. April 1946. No previous titer.

Rh	Mother	Father	5 yr.	Miscarriage	20 mo. (Pt.)
−	+	+		3 mos.	+ living and well

Screen test Agglutination with 4 Rh negative bloods
In albumin 1:1
In saline 1:1
Incomplete or blocking 1:1

3. R. W. May 1946. No anti-Rh

Rh	Mother	Father	19 mo. (pt.) (1st pregnancy)
−	+	+	Living and well

No anti-Rh agglutination or incomplete antibodies present.

4. D. P. August 1946. Anti-Rh negative 1:128

Rh	Mother	Father	16 mo. (pt.) (1st pregnancy)
−	+	+	living and well

Screen test Agglutination with 4 Rh positive bloods
In albumin 1:1–1:8
In saline 0
Incomplete or blocking 0

The mother had been sensitized 3 years previously by 3 transfusions.

5. N. P. August 1946. No previous titer

Rh	Mother	Father	Miscarriage	16 mo. (pt.)
−	+		7 mos.	+ living and well

Screen test Agglutination with 4 Rh positive bloods
In albumin 1:1–1:2
In saline 0
Incomplete or blocking 0

6. R. V. September 1946. No previous titer

Rh	Mother	Father	15 mo. (pt.)
+	+	−	

The child is still receiving Rh negative blood transfusions for Mediterranean anemia.

7. J. L. October 1946. No previous titer.

Rh	Mother	Father	Miscarriage	14 mo. (pt.)
	−	+	3 mos.	+ living and well

Screen test	Agglutination with 4 bloods
In albumin	1:1–1:2
In saline	0
Incomplete or blocking	1:1–1:2

8. D. S. November 1946. Anti-Rh 1:64

Rh	Mother	Father	9 yr.	1 died	13 mos. (pt.)
	−	0	+	E. F.	+ living and well.

Screen test	Agglutination with 4 Rh positive bloods
In albumin	1:1–1:2
In saline	0
Incomplete or blocking	1:1–1:2

9. K. S. August 1945. No previous titer

Rh	Mother	Father	2 stillborn	29 mo. (pt.)	8 mo.
	−	+		+	−

Screen test	Agglutination with 4 Rh positive bloods
In albumin	1:1–1:8
In saline	0
Incomplete or blocking	1:1

DISCUSSION

We have reviewed 66 cases diagnosed as erythroblastosis in Children's Hospital from January 1944 through September 1947. A review of the charts impressed us with the necessity of obtaining an antibody titer on the mother or infant to substantiate the clinical impression of erythroblastosis when the mother is Rh negative. Rh typing of the infant at birth can be misleading since the mother's antibodies may coat the infant's Rh positive cells, causing them to type Rh negative.

One case with an Rh negative mother and an Rh positive infant later proved to be a case of congenital syphilis. Two cases, in which the mother was Rh negative and the infant Rh positive, later came to autopsy where no evidence of erythroblastosis was found.

To complicate the picture further, one mother who was followed throughout pregnancy developed incomplete or blocking antibodies to 1:8 which, however, were not demonstrable shortly after delivery and had the previous examinations not been done, the diagnosis might have been missed.

In our cases, no correlation could be found between the height of the antibody titer and the clinical course of the disease in regard to the onset of symptoms, number of transfusions given, degree of anemia, presence of

erythroblastosis, length of hospital stay or whether the patient lived or died.

Five of the twenty-eight patients had slight or no jaundice. However, the red count in these 5 cases ranged from 1.9 to 4.0 million cells with an average of 2.8 million. There was one death in this group.

How then should an Rh negative woman be handled? In the first place the appearance of antibodies in a previously unsensitized Rh negative woman should be noted and followed by serial agglutinin titers during her pregnancy. Since the treatment of the complication, if it develops, will be handled at least in part by the pediatrician, it would appear to be advantageous for him to be advised in advance of the mother's past history and agglutinin titer and to be present at the delivery. It should be the joint responsibility of the obstetrician and pediatrician to determine whether or not an exchange transfusion is indicated. This procedure, in experienced hands, has been shown to be technically safe and is believed to offer the infant a better chance of survival, particularly if the mother has previously lost one or more children due to erythroblastosis. The decision to perform a replacement transfusion will have to be made at the time of delivery, taking into consideration the previous history of the mother, the rise of antibody titer and the examination of the infant and his blood, with demonstration of sensitization of the fetal cells by use of one of the developing tests such as mentioned by Davidsohn and Levine. Secondly, Rh negative blood should be used even though the infants are Rh positive. Third: These infants should not be breast fed. Fourth: Supportive therapy such as oxygen, stimulants, maintenance of fluid balance and protection of the liver if possible are necessary to complete the treatment of erythroblastosis fetalis.

Adrian Recinos, Jr., M.D.: In the series of 28 patients reviewed there were eight deaths. Postmortem examination was performed in seven. All were white; five were male and two female. Five of the seven cases apparently died in the hemolytic phase of the disease, the ages varying from 36 hours to five days. The other two died at 35 and 43 days respectively, one with biliary cirrhosis of the liver attributed to obstruction of the biliary ducts with impacted blood pigment, the other in acidosis and dehydration due to diarrhea.

Four of the five patients who died in the first five days of life presented typical pathological changes of erythroblastosis fetalis. The skin and sclerae were deeply icteric. The liver and spleen were markedly enlarged and histologically revealed congestion, excessive pigment deposition and abundant areas of erythropoiesis. Foci of nucleated red cells were found in some of the other organs. In two cases there was superficial edema and

fluid in the serous cavities. One of these was a typical "hydrops" type and died 36 hours after birth. The fifth case was the infant who received a "replacement" transfusion and died suddenly without satisfactory explanation at the age of three days. This patient had never been anemic and slight icterus was first noted on the day of death. At autopsy there was moderate icterus and slight edema. Yellowish pigment stained the basal ganglia and cortex of the brain. The liver and spleen were normal in size. Histologically, the spleen was essentially normal but the liver was congested, there were numerous foci of erythroblasts, and there was evidence of early cirrhosis. The absence of hepatosplenomegaly was the only variation from the usual pathological picture and can be explained by the absence of anemia. A direct cause of death could not be definitely established. The brain was examined in three of five cases who died in the first five days of life. Kernicterus was present and marked in all three. In each case pigmentation was present in other parts of the brain as well as in the basal ganglia. Kernicterus was absent and there were no other changes in the brains of the two patients who died after the fifth week.

Not included in this series are two infants who came to necropsy and may be considered possible or probable cases of erythroblastosis. They deserve mention because they illustrate some of the difficulties in making a postmortem diagnosis. The first of these was a six day old colored premature infant weighing 1740 grams. The mother was Rh negative and the infant Rh positive. No anti-Rh determinations were made. Jaundice appeared on the third day and the patient died after several episodes of respiratory distress on the sixth day of life. Postmortem examination revealed marked icterus, no edema, slight yellowish areas in the roof of the fourth ventricle and insula, a huge liver, and a grossly normal spleen. Microscopically, the spleen was not remarkable and small scattered areas of erythropoiesis were found in the liver. The lungs were atelectatic. Since it is known that erythropoiesis is a normal finding in the liver of premature infants, these changes can not be considered conclusive for the diagnosis of erythroblastosis fetalis.

The other case was a 13 day old Rh positive white infant. The mother was Rh negative and had had five previous pregnancies without viable issue. There were no anti-Rh determinations. Jaundice was present at birth and gradually deepened until death on the 13th day. Postmortem examination revealed marked icterus and hepatosplenomegaly. Microscopically, the liver and spleen were congested and contained abundant pigment deposits, and miliary abscesses were observed in the lungs. It was felt that a septicemia could account for these changes and for this reason a diagnosis of erythroblastosis fetalis could not be definitely established.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.
Assisted by: Harold W. Bischoff, M.D.
Adrian Recinos, Jr., M.D.
By Invitation: Richard Jackson, M.D.

Harold W. Bischoff, M.D.

This one month old white female was admitted to Children's Hospital on September 5th with the chief complaint of fretfulness and abdominal distention of 24 hours duration.

The day before entry the child sneezed; however, there was no other evidence of an upper respiratory infection. From the onset of the present illness the child had been irritable, not in apparent great pain but fussy, uncomfortable and crying more than usual. The morning of hospital entry the child had one bowel movement. There was no blood in this movement. The infant had two enemata during the day and 10 drops of paregoric because of apparent abdominal discomfort. Practically no food was accepted by the child during the day before entry. About four to five hours before admission the infant started to vomit and continued until one and one half hours after entry when Wangensteen suction was started. The vomitus eventually became yellow, mushy and sour. Abdominal distention at the time of admission was marked.

The family, birth and past histories were non-contributory.

Physical examination on entry revealed a markedly distended, month-old white female with mottled red skin and miliaria on the cheeks. The child frequently vomited a yellow, sour vomitus. Other than abdominal and rectal examination, the rest of the physical examination was normal. The abdomen was markedly distended, tight and drum-like. Tympany was reduced suggesting gas under pressure. Peristalsis was not heard. Palpation was of little help because of the distention. A rectal examination showed an "empty space" in the rectum. There was no blood on the examining finger. Sometime later bloody fluid (not mucoid) was passed.

A flat x-ray film of the abdomen in the upright position revealed the presence of several fluid levels in the small bowel, and probably some in the large bowel in the mid-descending region. There was a uniform opacity over the lower two-thirds of the abdomen.

Blood count on entry showed 11.0 grams of hemoglobin with 3.4 million red cells and 5,100 white blood cells with a normal differential.

A laparotomy was performed three hours after entry. Post-operatively the child did poorly and died approximately 6 hours after operation, 11

hours and 15 minutes after hospital entry and approximately 35 hours after the onset of symptoms.

DISCUSSION

Richard Jackson, M.D.: The acute abdomen in a month old infant is always an interesting problem. In this age group it is practically never inflammatory in origin. Acute appendicitis under two years is rare. The cause of an acute abdomen in an infant one month of age is usually due to congenital anomalies. There are not many other conditions that can cause an acute abdomen in a child of this age.

A child with pyloric stenosis usually reaches the surgeon 4 to 6 weeks after birth but the symptoms often start 2 to 3 weeks after birth. However, the clinical picture is not that of a pyloric stenosis and we can eliminate that as a possible diagnosis in this case.

Ninety-eight per cent of intestinal atresias reach the surgeon within the first few days of life and usually within the first 24 hours postpartum. This condition leads to vomiting and distention soon after the first food has been given. We can eliminate intestinal atresia simply on the basis of the history.

The next anomaly is intestinal stenosis. In this condition we do not get complete closure of the gastro-intestinal tract but there is usually a marked narrowing. Of Ladd and Gross' series, 10 out of 21 came to operation during the first week of life. The oldest case reported by Ladd and Gross was 9 years of age. If the child passes the first week of life, one usually finds a gradual onset of symptoms. Here we have a rapid onset of symptoms within 24 hours with distention and vomiting. This consitutes rather suggestive evidence against a diagnosis of intestinal stenosis.

Next we should consider a duplication of the gastro-intestinal tract. With a duplication of the gastro-intestinal tract there is a slow increase in the size of the duplicated bowel. An abdominal mass may have been noticed for weeks. There are no symptoms until there is encroachment upon the lumen of the bowel. There may be bloody stools. Quoting again from Ladd and Gross, the youngest patient was 2 weeks and the oldest 9 years of age. In all of these the first sign was an abdominal mass. All were followed for months, and all produced pressure symptoms. This clinical picture does not fit the case under discussion.

The next condition is intussusception. In one series of 480 cases there was only one child one month old. Approximately 75% of cases take place between 3 to 11 months of age. Intussusception is extremely rare in an infant 1 month of age or younger; however, it cannot be ruled out on age alone. I do not believe I have ever seen a case which the house staff did not diagnose before surgery. In the typical case the child screams with pain. Between pains the child plays and is quite comfortable. There is

no history of that here. Almost all of these children reach the hospital with a soft abdomen and a mass can be felt. In most cases we get the "currant-jelly" on the enema-tube or examining finger. Since this is a reliable history I feel we are justified in ruling out intussusception.

Another entity to consider is that of a developmental cyst, omental or mesenteric. These are due to a developmental anomaly of the lymphatic system. A mesenteric cyst is produced due to failure of the lymphatic tissue to communicate with the vascular system. These cysts may reach the size of a large grapefruit. In the infant, the cyst usually is felt before pressure symptoms arise. Here again the history is so short and rapid in onset that I feel this is not the primary pathology. On x-ray the fluid does not appear to me to be encapsulated, but seems to be free. This does not fit in with a mesenteric cyst.

Now we come to volvulus. This is a rather common condition seen early in life and is frequently associated with mal-rotation of the gastro-intestinal tract. It may be associated with a poor attachment of the mesentery posteriorly. Volvulus differs from intussusception in its history. The patient rarely gives a history of crying out with severe pain but one obtains a history of mild abdominal discomfort that may easily be overlooked for many hours. The diagnosis is much more difficult. What part of the intestinal tract is involved I don't know. I should say at this point that volvulus does not have to be associated with a poor attachment of the mesentery.

The history points to some obstruction of the gastro-intestinal tract and on admission the abdomen was silent. If irreversible changes in the gastro-intestinal tract have occurred due to impairment of the blood supply, the abdomen will be silent. The prognosis is much better if there is peristalsis.

So far as I can see, this case represents a volvulus and the silent abdomen means circulating changes have taken place. As a second choice I should say we should consider an internal hernia at the ligament of Treitz into the lesser peritoneal cavity although this is extremely rare.

PATHOLOGICAL DISCUSSION

E. Clarence Rice, M.D.: I would like to congratulate Dr. Jackson on the way he worked out his diagnosis. I concur on his comments on the frequency of occurrence of these conditions. The cysts, mesenteric and omental, are the most common.

This turned out to be an ovarian cyst about which a volvulus occurred. When one considers the size of the structures and the delicacy of the tissues involved, it is easy to see how difficult it must be for the surgeon at the operating table.

When the abdomen of this child was opened at necropsy, there was a gush of bloody fluid and all of the peritoneal structures were dark.

I will read the autopsy findings as they are here:

"*Gastro-Intestinal Tract:* The stomach is normal. The duodenum is somewhat distended but has a normal color. The small bowel from the jejunum to the ileocecal valve is distended, has a purple, hemorrhagic transparent color as does its adjoining mesentery with the lymph nodes. This area corresponds to the distribution of the superior mesenteric artery and is distended with gas and hemorrhagic mucus. The mucosa is shaggy and a red purple color. The appendix is about one centimeter long and of normal color. The colon, sigmoid and rectum have a normal color. The lumen is patent but empty. There is a ligature at the pelvic attachment of the infundibulo-pelvic ovarian ligament which is adjacent to the junction of the descending colon and sigmoid colon. This gave the erroneous impression at operation that the cyst removed was mesenteric.

Generative Organs: The left ovary has been removed. There is a ligature around the fallopian tube and adjacent broad ligament and the pelvic origin of the infundibulo-pelvic ligament which gives a very close approximation to the left horn of the uterus and junction of descending with sigmoid colon. The right ovary contains a hemorrhagic, purple cyst about 0.5 cm. in diameter; the fallopian tube and broad ligament appear normal.

The cyst removed from the left ovary was "kidney-colored", cystic in nature, weighed 35 grams and measured 4 x 6 cm."

So far as I know this is the first infant at Children's Hospital to have such a combination of findings. We see ovarian cysts in newborn babies but they are usually not more than a centimeter in size.